10

Methodology

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10.1 Planning and Feasibility Study
10.2 Questionnaire
10.3 The Team
10.4 Study criteria
10.5 Examination sites
10.6 Clinical Methodology
10.7 Data management and parameters
10.8 Definitions
10.9 Additional Studies

10.1 Planning / Feasibility Study

The survey was preceded by a feasibility study to establish the need and viability of conducting such a major work. Information was gathered from social services, school headmasters at blind schools and institutes and their permission gained. A protocol was prepared and submitted to St John Ophthalmic Hospital (SJOH) and the Order of St John in London for ethical approval. Final approval was finalised with the social services, schools and civil authority.

Data on the schools, their locations in both the West Bank (WB) and the Gaza Strip (GS) were gathered.

The final stage was personnel recruitment and funding. The team comprised members of the Out-Reach Team on a voluntary basis.

Registers were created for various data. Recruited cases were given a number which combined the provisional diagnosis and a serial numbers for the family and siblings who fulfilled the criteria of the study.

Protocol forms were designed using Amstrad PCW 8256 by the author. These included; a form for family details; ophthalmic examination form, genealogy tree form, and a questionnaire in both Arabic and English.

An instruction booklet with guidelines including instruments, drops, medications, etc. to carry in the mobile eye unit, protocols to carry, and protocols for visual acuity assessment and colour vision testing by nurses were prepared, An office was allocated at the hospital for the use of the administrative team etc. [1]

10.2 Questionnaire

Questionnaires were devised in Arabic to assist the team and the resident social workers to obtain information from the parents about their children and siblings prior to the home visit and this information was then transferred to the equivalent questionnaires in English by the trained bilingual social worker.

Specially prepared protocol forms were designed for family history and sibling information including pre-, peri-, postnatal and adolescent histories.

10.3 The Team

The team included a dedicated Blind Schools Survey team and the Outreach Programme team. The BSS team comprised a full-time research assistant/social worker (from June 1986); one part-time personal secretary/research assistant (from October 1986); and one full time junior research assistant who was later trained as an electrodiagnostic assistant technician (from March 1987). The Outreach team comprised a driver / paramedic; 1 permanent outreach nurse;

and an additional 1 to 2 non-permanent nurses. Visiting orthoptists joined the team when time permitted; alternatively patients were sent to SJOH for orthoptic assessment

10.4 Study Criteria

The criteria for inclusion in the study were childhood onset bilateral visual impairment in the population of the West Bank (WB) and Gaza Strip (GS).

10.5 Examination Sites

Each of the blind institutions were visited by the Blind School Survey (BSS) Team using the 'Outreach' Mobile Eye Unit facility of SJOH.

When circumstances prevented families from attending the school/institute or the hospital, or extensive work was required, (particularly where several members of the family were involved), the team visited the family in their home (Plate 9). This approach was used extensively when collecting genealogy information from the family elders, and relied on the oral tradition. The work was often done with the collaboration of the local social services workers and the village "Mukhtar" (village chief of mayor of a village).

Patients and Schools

All pupils, affected siblings, trainees, blind teachers, and younger children on the school's waiting lists were examined. Outpatients meeting the criteria were also included in the study. Parents and siblings of the affected patients were also screened. The study lasted from October 1985 to September 1987. (Chapter 10 Appendix)

At the completion of the survey, 709 cases were enrolled, 40 of them were later excluded from the study for not fulfilling the study criteria of childhood onset bilateral visual impairment in the WB and GS, concluding with 699 cases.

Blind institutes in the WB comprised 3 primary schools for girls funded by local and international charities and one school for boys which provided both primary and secondary and was funded by the Department of Education. The former three also accommodated very young boys. The remaining two institutions were residential for all disabilities and housed older students in higher education. The primary school in Nablus also contained a workshop for older girls who did not proceed to higher education. All the schools in the WB followed the Jordanian curriculum. Boys progressing to university education who lived away from the university sites either stayed at the residential homes in the WB or moved to Jordan. A few children from affluent families were sent to Europe and America for further education.

In the GS, there was one mixed primary school funded by UNRWA which also contained a workshop for older girls. The school was non-residential and children who progressed to secondary education joined the sighted school in the GS. Students from Gaza who joined WB universities resided in residential homes in the WB. Gaza schools followed the Egyptian curriculum.

10.6 Clinical Methodology

Pre, Peri and Post-Natal Histories

Detailed prenatal, perinatal and postnatal data, together with full ophthalmic, medical and genetic histories were taken from the patients and/or their parents.

Demographic/ Genealogy Data

Detailed data was collected where possible of the patients, their siblings and half siblings and their parents, including details of the mother's family and the extended family known locally as "Hamoula".

Genetic Protocol

Full marriage details were sought including the degree of consanguinity as far back as was known. Pedigree charts were drawn from the data available and frequently checked and revised to eradicate errors, especially in the large pedigrees. Patients were placed on the chart and other family members were plotted. The exact sequence and gender of births, including any

prenatal and postnatal death was observed with as much accuracy as possible. Lengthy family interviews delineating all family relationships, reliant upon the strong oral tradition of family genealogy held by family members and the elders, were carried out to establish relatedness and identify genetic families. The degree and extent of cousin marriage was recorded and traced as far back as possible on the pedigree chart. Where it was certain that marriage was from the same family, but the exact relationship could not be ascertained with certainty, either from history or on the family chart, it was recorded as 'same family'. If consanguinity was absent, the geographic origin of the parents was investigated ie whether they came from the same village, town or district etc. 'No relationship' was marked on the family chart when this was stated and confirmed.

Clinical History

In addition to routine clinical histories emphasis, in particular with retinal dystrophies, was given to the presenting symptoms or features and the time of their onset, the course of the disease and the presence and/or absence of photophobia and night blindness.

Clinical Examination

Full ophthalmic and, when relevant systemic examinations, included visual acuities, cycloplegic refraction, orthoptic examination, slit-lamp biomicroscopy and fundoscopy with mydriasis was performed the Author.

Ophthalmic tests including psychophysical and electrophysiological tests were performed. Visual field assessment was carried out by confrontation method but, when possible, Goldman perimetry was performed at the base hospital. Electrophysiological tests included full-field electroretinography (ERG), electro-oculography (EOG) and visual evoked potentials (VEP). The full protocol of elctroretinography protocol is described elsewhere.[(2), (3), (4)]

Apart from the cases where information was extracted from SJOH casenotes at the very end of the study, all patients were examined by the author. Rare and interesting cases were seen at the weekly clinical meeting at the hospital.

When further assessment was required, e.g. examinations under anaesthesia, fundus photography and fundus fluorescein angiography or orthoptic follow-up, patients were seen at the base hospital. Electrodiagnostic tests were performed three months prior to the end of the survey when facilities became available.

Dental work was initially carried out by a local volunteer dentist who participated in a one-day fieldwork with the author.

Patients who required medical opinion were referred to either St Joseph's Hospital (Neurology and ENT) or Al-Makasid Hospital for medical conditions. Surgical opinion was provided for this research on the SJOH site by Dr Peter Qumri, Consultant Surgeon at Beit Jala Hospital.

Assessment of Visual Functions

Visual acuities: both unaided and best corrected, were measured using Sheridan Gardiner test charts and, for the very young, cake decorations. When necessary, E-test chart was used.

Colour vision: Ishihara Pseudo-isochromatic Plates, City University Plates, text reference for colour to assess gross colour and, in the severely visually impaired, Kodak red filter C-25, and the green and blue filters of the slit lamp.

Visual fields: Performed using confrontation method, portable Bjerrum screen and Goldman Perimetry when further examination at SJOH needed.

Electrodiagnostic assessment

This was preceded by the setting up of normal values and staff training carried out initially by Mr Chris Hogg, Senior Technician, at the Electrodiagnostic Unit, Moorfields Eye Hospital, London.

Photography and fluorescein angiography:

Performed when applicable at SJOH.

Further investigations: biochemical tests and blood samples for molecular studies were taken

from patients with suspected metabolic disorders and for molecular biological studies.

Table 10.1 WHO categories of visual acuities

Category	Description	Visual Acuities
'1'	No impairment (NVI)	6/18 or better
'2'	Visual impairment (VI)	<6/18 to 6/60
'3'	Severe visual impairment (SVI)	<6/60 to 3/60
'4'	Blindness (BL)	<3/60 to LP
'5'	Blindness (BL)	NLP
Patients on whom reliable assessment of vision was not possible have been categorised by the WHO under categories '6' and '7'. Unavailable data was included under a new category 8.		
'6'	Believed to be sighted but with some visual impairment	
'7'	Believed to have significant visual disability or to be blind.	
'8'	No reading is available, such as unable to examine, lost data, etc.	

10.7 Data Management and Parameters

Preliminary Clinical Classifications
Conditions were initially classified clinically. After the data was compiled on a database, WHO modified categories including visual acuities, anatomical and aetiological classifications were applied. [5]

Database
Data was compiled on the special protocol forms and later transferred to a database (Microsoft Access). A spreadsheet (Microsoft Excel) was used for statistical analysis, tables, calculation and graphs.

Visual Acuities
Visual acuity levels were analysed according to WHO/PBL categories (WHO/PBL Eye Examination Record for Children with Blindness and Low Vision, Coding Instruction), [5], [6] The same protocol was applied for anatomical and clinical classification including where there was a difference in the anatomical diagnoses between each eye in which case the diagnosis of the Best Seeing Eye was used. For the visual acuities, WHO categories for visual acuities were used in the analysis. (Table 11-1)

Age group used in statistics
Epidemiological figures on childhood blindness adhered to children at or below the age of 16 years at the time of examination. Many of these had passed the age of 18 at the completion of the study which lasted 2 years.

Statistical Analysis and Prevalence
Excel spreadsheet were used. Prevalence figures used the established formulae. [7]

Formulae of blindness and other conditions was based on under 16 years old population of 562 300, 322 000, and 884 400 in the WB, GS, and both regions combined at the time of the survey and were based on the Palestinian Central Bureau of Statistics 1994, Department of Users Services, Demography of the Palestinian Population in the West Bank and Gaza Strip, Current Status Report Series 1. Ramalla-West Bank (Appendix 1).

10.8 Definitions of Childhood and Childhood Blindness

Childhood is defined as 0-16 years inclusive. Childhood blinding conditions are a group of diseases and conditions occurring in childhood or early adolescence, which, if left untreated, result in blindness or severe visual impairment that are likely to be untreatable later in life. **[8] [9]**

WHO categories of visual impairment (VI) (Table 10.1) defines blindness (BL) as a corrected visual acuity in the better eye of less than 3/60, and severe visual impairment (SVI) as corrected visual acuity in the better eye of less than 6/60 but equal to, or better than 3/60. **[10]**

10.9 Additions to the Survey

Retrospective review of casenotes

Before the completion of the study and in order to provide more insight into changing patterns in the under-5's and more reliable prevalence data, all casenotes of patients under the age 19 were reviewed. Patients who fulfilled the criteria of childhood onset bilateral visual impairment were added to the study and analysed.

Two additional elements were later added to the protocol. These were:

Red-Tinted Filter (Glasses)

The efficacy of red-tinted glasses (filter) in providing symptomatic relief and alleviating photophobia to photophobic patients and comparing them to commercially available types of tinted sun glasses. This off shoot of the study focused on cases with photophobia, in particular cone disorders and albinism. (Figure 10.1)

Educational Performance and skills

The second added parameter was the educational performance of pupils using school marks as a crude parameter to assess their educational performance and abilities in relation to the type of pathology they have. A randomly selected sample in proportion to the size of cohort in each condition was planned. This was not possible to achieve fully as a bias of the schools towards their better achievers resulted in under representation of some conditions.

Figure 10.1 Red filter glasses used in the trial.

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Appendix Chapter's 10 Cases and schools and institutes in the survey of childhood onset visual impairment in the West Bank and Gaza Strip.

Institute/Residence	Pupils[a]	Residents	W/L[b]	Relatives	Total	Type of Institute
West Bank						
Al-Ala'iyeh (Bethlehem)	46	-	-	13	59	Boys, primary / secondary
Al-Bireh (Ramalla)	57	7	4	24	92	Girls, primary, boarding
Al-Shurooq (Beit Hanina)	30	20	-	7	57	Girls primary, boarding
House of Hope (Beit Jala)	-	28	-	2	30	Residential home
House of Light (Beit Jala)	-	1	-	0	1	Residential home
Al Nour (Nablus)						
School	5	-	-	6	11	Small girls primary
Training Centre	9	-	-	-	9	Training workshop
Gaza Strip						
UNRWA (Gaza City)						
School	77	-	21	91	189	Mixed primary
Training workshop	14	-	-	-	14	
Residential / Multiple Handicapps homes	4	-	-	-	4	Residential/multiple
Subtotal recruits via schools	**242** [c]	**56**	**25**	**143**	**466**	
Out-patients (OP) and home visits [c]	202	-	-	1	203	All age groups
Total analysed [d]					**669**	

[a] includes pre-school inhabitants and 6 in higher education elsewhere.
[b] Pre school children on the schools' waiting list.
[c] Also includes relatives and some 85 cases from the OP case notes.
[d] This is derived after excluding 40 cases for not fulfilling criteria: 40 OPs and 14 seen at Gaza school.